## **Clinical Quiz**

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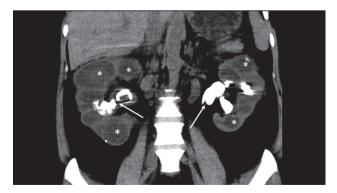
#### Adult female with acute renal failure and weight loss

#### **Clinical Presentation**

A 46-year-old woman presented with a 6-month history of anorexia and loss of weight. This was associated with a 2-month history of cloudy urine. The only significant past medical history was a previous ectopic pregnancy. On examination, she was hypertensive with a blood pressure of 196/102 mm Hg. The patient was afebrile with a normal pulse and respiratory rate. Abdominal examination revealed non-tender ballotable kidneys bilaterally. Her renal function was impaired with a urea of 26.2 mmol/L (normal range: 73.4 mg/dL) and a serum creatinine of 790 umol/L (normal range: 8.94 mg/dL). Urine analysis revealed proteinuria, hematuria, and a leucocyte count of >1000/HPF. Blood count showed a normocytic anemia of 7.7 g/dl and a raised white blood cell count of 13.0 x 10<sup>9</sup>/L. She was rehydrated and treated with antibiotics. Renal ultrasound showed dilated calyces bilaterally with bilateral renal calculi. An initial diagnosis of obstructive uropathy was made and bilateral retrograde pyelogram (RPG) and stenting was carried out. Plain radiograph (Figure 1) and a plain CT of the abdomen (Figure 2) were carried out to evaluate the cause of the urinary tract obstruction. As the renal function remained severely impaired in spite of this procedure, bilateral nephrostomy was performed. Culture of pus aspirated from the renal calyces yielded Proteus mirabilis and Pseudomonas aeuroginosa. The urine culture was negative. Following discussions in multiple multi-disciplinary meetings, it was decided that she requires a bilateral nephrectomy, which was carried out with no immediate complications. Hispathological evaluation of the kidneys revealed acute inflammatory exudates and mixed inflammatory cells including neutrophils, lymphocytes, plasma cells, multi-nucleated giant cells, and abundant foamy histiocytes forming xanthogranulomas. She was discharged 10 days after surgery with regular dialysis.



**Figure 1** - Plain film of the abdomen showing bilateral staghorn calculi (white arrows).



**Figure 2 -** Coronal CT image showing bilaterally enlarged kidneys with bilateral staghorn calculi (white arrows), dilated calyces (asterisks) and paradoxical contracted renal pelves.



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## **Questions**

- 1. What are the findings on plain film and CT of the abdomen.
  - 2. What radiological sign is seen on the CT?
    - 3. What is the diagnosis?

## **Answers**

- 1. The plain radiograph of the abdomen shows bilateral staghorn calculi with enlarged renal silhouette bilaterally. The CT shows dilatation of the renal calyces bilaterally with paradoxical contraction of the renal pelves.
- 2. Dilatation of the renal calyces with paradoxical renal pelvis contraction gives the bilateral "bear paw" sign.
- 3. Xanthogranulomatous pyelonephritis.

## **Discussion**

Xanthogranulomatous pyelonephritis (XGP) is an uncommon form of bacterial pyelonephritis with an incidence of 1.4 per population of 100 000 per year. Almost all cases of XGP are unilateral. Bilateral involvement is extremely rare. Chronic obstruction of the urinary tract leads to destructive replacement of the renal parenchyma by granulomas, abscesses, and lipid laden macrophages, which is the hallmark of XGP. Diffuse XGP accounts for 90% of cases and focal XGP accounts for the remaining 10%. The typical CT findings in diffuse XGP are of dilated calyces with paradoxical contracted renal pelvis giving a 'bear paw' sign, with the dilated calyces resembling the toe pads of a bear's paw. Focal XGP exhibits similar changes confined to an upper, or lower pole calyx. Although the CT findings highlighted in our case are typical, the CT findings in XGP have a wide range of findings such as diffuse renal enlargement, renal atrophy, hydronephrosis, focal masses and air within the urinary tract. Due to the wide range of CT findings, differentiating XGP from renal tumors and abscesses on imaging can be challenging preoperatively. The definitive treatment of diffuse XGP is complete nephrectomy and resection of any involved tissue. Partial nephrectomy has only been shown to be of benefit in focal XGP.

In conclusion, XGP is an uncommon form of bacterial pyelonephritis with a wide range of CT findings. The bear paw sign describes the classic appearance of dilated calyces with relative sparing of the renal pelvis. Awareness of the imaging signs in XGP can lead to earlier diagnosis and appropriate treatment.

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#### References

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#### **Case Reports**

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